CASE REPORT

Juvenile nasopharyngeal angiofibroma in a 20 year old Nigerian male

Amali Adekwu[1], Jacob Ausha[2], Terna Gav[3], Raymond A Vhriterhire[4]

Abstract

Background: Juvenile Nasopharyngeal angiofibroma is a rare benign tumour of the nasopharynx that occurs predominantly in adolescent males in the second decade of life. This paper presents misdiagnosis of a 20 year old male with Juvenile nasopharyngeal angiofibroma (JNA).
Methods: The case record of a 20year old male who presented with recurrent spontaneous profuse epistaxis, progressive nasal obstruction, hyponasality and conductive hearing loss with mass in the post nasal space was reviewed.
Results: Examination under anaesthesia revealed a firm, purplish, hemorrhagic smooth tumour filling the nasopharynx. He had a trans-palatal excisional biopsy for which histology confirmed angiofibroma.
Conclusion: Male juvenile patients presenting with recurrent epistaxis need early referral to the Otorhinolaryngologist for further evaluation.

Keywords: Epistaxis, Angiofibroma,

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Introduction

Juvenile Nasopharyngeal Angiofibroma (JNA), is a rare tumour of the nasopharynx of unknown aetiology and predominantly seen in adolescent males in the second decade of life[1,2]. It accounts for 0.05% of all head and neck tumours and occurs at a frequency of 1:5000 to 1:60,000 in Otorhinolaryngology patients[2]. The diagnosis is mostly based on clinical presentation as biopsy is attended with profuse bleeding and is therefore avoided[2,3]. Patients with JNA manifest with profuse and recurrent epistaxis, progressive nasal obstruction, denasal speech, conductive hearing loss, serous otitis media and mass in the nasopharynx [1,2]. This presentation demonstrates misdiagnosis of an adolescent male with a rare tumor thought to be hypertensive epistaxis.

Case Report

YD is 20 year old male who was referred to the Otorhinolaryngologist by the General Practitioner on account of worsening recurrent, profuse, epistaxis with dizzy spells after managing him for three years for epistaxis secondary to hypertension. Patient had received three pints of blood transfusions in the past one year. Additional history obtained included progressive bilateral nasal obstruction, impaired hearing and denasal speech of one year duration. Examination revealed a young man who was mouth breathing, moderately pale but not in any distress. Anterior rhinoscopy revealed normal nasal pyramid, absent airflow in both cavities but no mass seen in the nose. Oropharyngeal examination revealed bulging soft palate while the tonsils were not enlarged. There were no palpable neck nodes. Ear examination revealed bilateral mild conductive hearing loss evidenced by negative Rinne tests and pure tone averages of 38dB and 40dB for the right and left ears respectively with type B tympanograms in both ears. The chest and abdominal examinations were essentially normal.

Radiograph of the postnasal space showed soft tissue mass completely occluding the nasopharynx while paranasal sinus radiographs showed no maxillary antral involvement. CT scan of the paranasal sinuses and nasopharynx could not be done. A diagnosis of Juvenile Nasopharyngeal Angiofibroma was made. Patient was worked up for examination under anaesthesia (EUA) and Excisional biopsy. He had two pints of whole blood transfused before surgical intervention. The EUA findings were that of a non-pulsatile, firm, hemorrhagic purplish tumour filling the nasopharynx (Figure 1). Thereafter, he had transpalatal excisional biopsy of the tumour. Bleeding was controlled with the use of bipolar cautery and adrenaline pressure packs. Estimated blood loss was about 750mls. Patient received additional 2 pints of fresh whole blood intra- and post-operatively and his packed cell volume had remained within normal limit. A

[1]Department of Human Physiology, Faculty of Medical Sciences, University of Jos, Jos [2]Department of Surgery, Federal Medical Centre, Makurdi [3]Department of Surgery, Benue State University Teaching Hospital, Makurdi [4]Department of Histopathology, Benue State University Teaching Hospital, Makurdi

All correspondences to:
Dr Amali Adekwu
Department of Human Physiology, University of Jos, PMB 2084, Jos
E-mail:amaliadekwu@yahoo.com

posterior nasal pack was left in situ for 72 hours. Histology of the tumour confirmed angiofibroma (Figure 2).He was discharged on the 10th post-operative day and followed up in the out-patient clinic for 6 months without events.

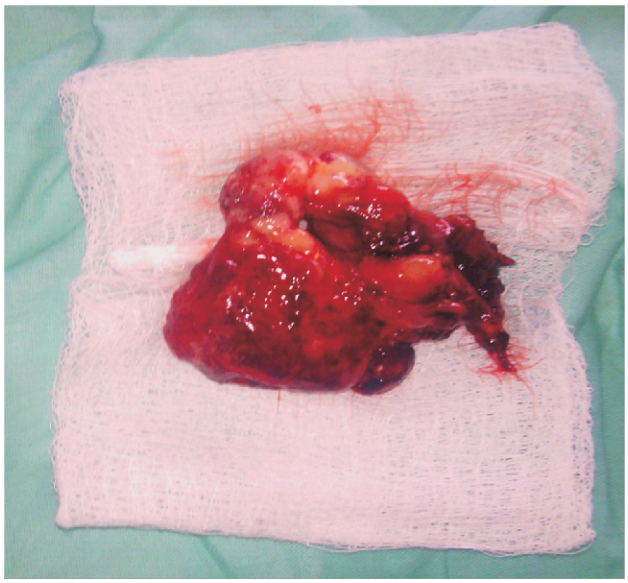

Figure 1. The gross image of the tumour removed from a juvenile male presenting with recurrent epistaxis

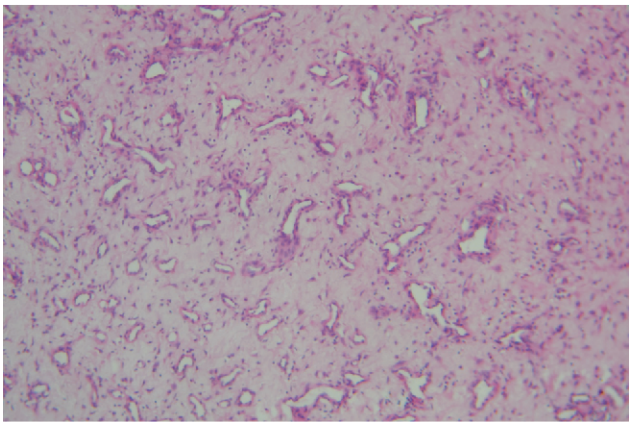

Figure 2. Histologically the tumour shows a fibrocollagenous stroma vascular spaces of different sizes which are round, stellate to staghorn in shape and are lined by endothelial cells (x 40 magnification).

Discussion

JNA, though a rare tumor is the commonest of all benign tumors of the nasopharynx[1]. The etiology of JNA is unknown; however it is thought to be testosterone dependent. Adolescent males with this tumor have harmartomatous nidus of vascular tissue in the nasopharynx which is activated to form angiofibroma, when male sex hormone appears[1,2]. Our index case is an adolescent male which agrees with this theory.

Clinical presentation of profuse, recurrent epistaxis with anemia, progressive nasal obstruction, hearing loss and mass in the nasopharynx were all demonstrated in this patient. Imaging studies, specifically a CT Scan and/or an MRI is necessary to determine the extent or staging of the tumor[3]. These were not done due to lack of funds.

There are various surgical approaches to the JNA depending on its origin and extensions which includes trans-palatine; trans-palatine and sub-labial (Sadana's procedure); extended lateral rhinotomy (via facial incision, or degloving approach); extended Denker's approach; intracranial-extracranial approach; infratemporal fossa approach and recently intranasal endoscopic approach [1,2,3].

Different authors reported varying degrees of success in the surgical management of JNA using these various options. Though our patient had a trans-palatal approach with good result, other authors like Blount et al[2], Boghani et al[4] and Mistry et al[5] advocated endoscopic resection which has less intra-operative blood loss, better cosmetic effect and lower recurrence rate. Also, the use of pre-operative embolization before the choice of any surgical approach will result in less blood loss as highlighted by Tyagi et al[6] and Mistry et al[5] in their separate studies. Our Centre lacks facilities and expertise for both embolization and endoscopic surgery. McAfee et al [7] in their study reported a high likelihood of cure after definitive radiotherapy for advanced and/or recurrent JNA with low risk of complication. But since our index case has not shown any symptom of recurrence, he was not recommended for radiotherapy.

This report highlights the need for early referral of recurrent epistaxis especially in adolescent males to the Otorhinolaryngologists for further evaluation as there may not be associated with 'hypertension' as was erroneously thought in this case.

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